OTS: 60-41,402

MAIN FILE

JPRS: 5600

21 September 1960

5600

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Distributed by:

OFFICE OF TECHNICAL SERVICES
U. S. DEPARTMENT OF COMMERCE
WASHINGTON 25, D. C.

Price: $0.50

U. S. JOINT PUBLICATIONS RESEARCH SERVICE
205 EAST 42nd STREET, SUITE 300
NEW YORK 17, N. Y.

JPRS: 5600
CSO: 4572-N

TREND IN SUBSTANTIATION OF THE DIAGNOSIS OF NEURO-ECTODERMAL TUMORS OF THE CEREBRUM

[Following is the translation of an article by M. Yu. Rapoport entitled Napravleniye v Obosnovanii Diagnoza Neyroektodermal'nykh Opukholey Bol'shikh Polusharly Mozga, (English version above), in Voprosy Neyrokhirurgii (Problems of Neurosurgery, Vol. XXIV, No. 3, Moscow, pages 14-19.]

Problems of active management and neurosurgical tactics in cerebral gliomas have been discussed in a contradictory manner and from different points of view.

In the investigation of gliomas it is now inexpedient to be limited to the principles of Cushing and Baily, who during the course of three decades have determined the trend of works of neurosurgeons, pathomorphologists and neuropathologists. New lines of pathological investigations of gliomas have been planned by works carried out in the USSR by L. I. Smirnov (L. I. Smirnov. Histogenesis, Histology and Topography of Brain Tumors. Part I, 1951; Part 2, 1959) and in Germany by Zülleh (K. J. Zülleh. Die Hirngeschwülste in biologischen und morphologischen Darstellung. 2. A. Leipzig, 1956).

The first problem of the clinician is that of giving a firm basis to the diagnosis of glioma with differentiation from tumors

of the meningovascular type. The local diagnosis within limits of an individual lobe or of tumor involvement of contiguous lobes which is determinable by means of neurological methods should be regarded as the basis of diagnostic analysis.

The next problem is the determination of the probable nature of the glioma and an evaluation of its maturity, a determination of its solidity or of the presence of cyst formation in it, a tendency to hemorrhages, circulatory disorders and necroses, to general or local edematous reactions.

Considerations of the apparently glial nature of the tumor, its location and extent and of the most probable structure of the glioma within limits possible for clinical examinations and analysis should be regarded as the main guiding points for further substantiation of a precise neurosurgical diagnosis of a cerebral glioma.

Neurosurgical practice has justified the distinction of three main groups of gliomatous growths: 1) cortical-subcortical, 2) subcortical in the area of the centrum ovale, 3) deep periventricular with involvement of the subcortex and its ganglia. During the course of tumor growth, naturally, transitions from one group to another are possible.

In the evaluation of the principal forms of location and growth of gliomas the need occurs for distinguishing, according to L. I. Smirnov, the following gliomas: 1) nodular, which to a certain degree are delimited, without features of infiltrative growth; 2) nodular with features of infiltrative growth; 3) diffuse tumor growth. The morphologically indicated division cannot be made with the necessary definition and surety; however, in the neurosurgical clinic the grounds for such attempts at differentiation are adequately weighty. This primarily deals with cortical-subcortical gliomas; to a lesser degree, subcortical gliomas in the centrum ovale, their mature forms, which not uncommonly occur with cyst formation, and sometimes a tendency toward loss of differentiation. All varieties of immature tumors, particularly with an initial periventricular growth as well as growth in the centrum ovale, stand in contradistinction to the forms mentioned.

Generalizations and conclusions made in the study of numerous patients with confirmed cerebral gliomas were supported by an extensive comprehensive investigation with the use of all the methods of clinical, laboratory, X-ray and electrophysiological analysis.

On the basis of varied clinical investigations, neurologically

and at the same time neurosurgically directed, the probable gliomatous structure of the tumor, its location and its relationship to the superficial cortical-subcortical areas, to the subcortex of the centrum ovale or to the deep periventricular areas of the subcortex can be determined by analyzing the sequence of development of the tumor syndrome and the characteristics of its course. The sequence of manifestation of local and general symptoms, their interrelationship in the dynamics of the tumor syndrome, the evaluation of the characteristics of the entire course create conditions for a preliminary evaluation of the probable nature of the glioma, the possibility of cyst-formation in the tumor, or the development of circulatory processes in it.

The indications, choice and sequence of application of various precision methods of neurosurgical examination depend on the group to which the given glioma belongs, its location, maturity, the degree of local and general cerebral reactions to the tumor growth. The stage of the disease, the nature of the earlier phases and the late periods, the nature of the diffuse general and secondary phenomena, including brain stem phenomena, are of great importance.

In the superficial cortical-subcortical gliomas with local

neurological syndrome, when the signs of increased pressure are slight and there are no brain stem symptoms, the importance of clear-cut manifestations of the focus of pathological electrical activity of a primary nature in phases where there are only slight or no signs of overt general cerebral disorders on the electroencephalogram (EEK) is great.

Spinal tap is contraindicated in the far advanced stages of the disease or in rapid cyst-formation processes or in the presence of circulatory disorders. Pathological changes are found in the spinal fluid in the case of neoplasms which have lost their differentiation or in case of neoplasms which are immature and in case the tumor ir in the direct vicinity of the meningeal spaces.

The method of arteriography is valuable for differentiating gliomas from arachnoidendotheliomas or angioreticulomas, and for the purpose of determining the degree of vascularization of the glioma.

During the period of absence of brain-stem displacement the pneumoencephalogram can confirm the conclusions based on the investigations mentioned above, and can assist in making the localization and size of the tumor more precise. Ventriculography

may be technically difficult to accomplish. The choice of the site of ventricular puncture, the evaluation of the data obtained from puncture and from the injection of air depend on the location of the tumor.

A special study of our own observations has shown that the cortical-subcortical gliomas can remain undiagnosed for a long time when only one of the precision methods is used, particularly, air contrast, and then they are handled incorrectly even under conditions of a many-sided clinical investigation.

The combination and sequence of methods of neurosurgical diagnostic precision significance acquire special importance specifically in the cortical-subcortical gliomas, which not uncommonly are nodular and which macroscopically are sufficiently clearly delimited, which give a good prognosis after radical removal (they require radical removal even in those cases where the operation was undertaken because of a suspicion of a meningioma).

We cannot overlook the manifestations of deterioration and cyst formation in these tumors and in addition we cannot overlook the fact that at the operation they can prove to be

infiltrative in some areas. Consistent, many-sided investigation directed by clinical observation is of decisive importance in the diagnosis and choice of surgical tactics. The method of prolonged observation is not suitable here.

Accurate diagnosis of gliomas of the subcortical type located in the centrum ovale has its own characteristics and difficulties. In many patients the neurological local syndromes are manifested in those cases with inadequate definiteness, and therefore, they have less topical-diagnostic value, particularly in the case of cysts within the tumors. Most often, complicated syndromes of subcortical involvement of two or even three adjacent lobes are observed, not uncommonly with indistinct individual elements. In substantiating the diagnosis the need arises for distinguishing neurological signs which make it possible to judge the area of early tumor involvement, the sequence of involvement of various lobes, the clinical expression of the direction of tumor growth, the characteristics inherent in cyst-formation.

Difficulties and errors in the neurological topical diagnosis are brought about by the manifestations of the dynamic localization of function and the possibility of topical variety in a

number of clinically related syndromes (disorders of the higher functions -- speech, praxis, orientation and complicated phenomena in the sensory sphere). The separation of symptoms of a primary nature from those of secondary nature and general nature is an inevitable condition. Electroencephalographically, the focus of pathological electrical activity is found with considerably less distinctness in the subcortical gliomas, and here usually it is only of regional-focal significance. With a marked increase in intracranial pressure the focus of pathological electrical activity becomes progressively more indefinite and it is very difficult to distinguish it from regional-focal manifestations of secondary nature. In the presence of malignancy of gliomas of the centrum ovale diffuse slow waves, which are different from those characteristic of general EEG changes during rapid cyst-formation in mature gliomas or in the far advanced stages are noticed early, even before the development of papilledema, against the background of a focus in the EEG

Any considerable changes in the spinal fluid are observed rarely in such cases, and then, in the less mature tumors, among those which are richly vascularized with a tendency

toward circulatory disorders.

Only in the cases of considerable blood-vessel-formation, particularly in the glioblastomas, can arteriography make clear the nature of the tumor. In mature oligodendrogliomas and astrocytomas not uncommonly only tension of the blood vessels in the area of the tumor is found by means of arteriography, sometimes with displacement of the deep arteries and veins and particularly with considerable congestion of these veins, as has been indicated by M. B. Kopylov. The use of angiography here should be considered necessary, and in cases where rich vascularization and circulatory disorders are suspected in the tumor, it should be considered indicated.

In substantiating a generalizing preoperative conclusion for gliomas of the centrum ovale a very important part is played by the air contrast examination. The use of ventriculography or encephalography in these cases depends on a number of conditions. The first of them is the convincing nature of the X-ray data, which is obtained only with adequately large quantities of the air introduced, which is not always possible or safe for the patient; second the performance of the operation directly after the air contrast examination. Hence, it is an obligatory requirement to

have a preliminary idea of what specifically should be suspected with the greatest probability: a glioblastoma of the malignant type or the presence of a more mature glioma and the possibility of considerable cyst formation in it.

The plan of operation is determined by the topical characteristics. Because of the localization of a considerable number of centrum gliomas of the brain in the subcortex, in the area of the ovale and the frequency of mature forms of tumors with cystic degeneration operative procedures on them give a good effect in the majority of patients.

The severity of reactions to air contrast in glioblastomas and in the late stages of growth of all gliomas requires the soundest possible foundation for its application. A comparison of the data of the neurological examination dynamically, the results of the EEG, of investigation of the spinal fluid and arteriography make it possible to gain an idea of the possibility of allowing the use of air contrast, which here should not uncommonly be considered indicated for making the location and volume of the tumor more precise, as well as for the surgical tactics.

In deep subcortical tumors of the periventricular area infiltrative forms not uncommonly predominate with frequent

involvement of subcortical ganglia, but delimited tumors are encountered also. These are considered inoperable by neurosurgeons. In the presence of indications, in these cases, recourse should be had to decompression operations and subsequent radiation therapy. This requires attentive differentiation of gliomas of this category. It is necessary to take into consideration the fact that the brain of patients with tumors in this localization gives severe reactions to diagnostic procedures, particularly to contrast procedures with the use of air.

Neurological syndromes which make it possible to suspect a deep tumor of the hemisphere with periventricular extension of it are made up of subcortical symptoms which manifest themselves differently depending on the topical factors which are frequently expressed with inadequate distinctness, not uncommonly on relatively early signs of involvement of subcortical ganglia and those characteristics of neurological symptomatology which are brought about by involvement or reactions of periventricular nature. The latter is expressed in the tendency of cerebral gliomas of this category to produce considerable local and general reactions in the form of edema and circulatory disorders

in the tumor and brain tissue with the threat of rapid development of inter-brain and mid-brain symptoms, to which pontobulbar symptoms are also added.

Signs of increase in intracranial pressure in such cases can be early, but can also be absent for a certain time despite the stormy clinical manifestations of focal and general cerebral disorders. This characteristic feature not uncommonly even in the early stage of the disease gives one a tendency to suspect a vascular disorder, and sometimes an inflammatory one, to which changes in the spinal or ventricular fluid noted can contribute fairly often.

Craniography frequently does not give any supporting data of diagnostic value.

The investigation of the spinal and ventricular fluids can assist the characterization of a periventricular tumor very much. The predominance of the protein content in the ventricular fluid, sometimes with inflammatory changes of the cell composition and in some cases elements of xanthochromia cause the clinician to be on special guard. The spinal and ventricular punctures should be made after the preliminary analysis of the symptoms and a discussion of the indications for these punctures. With a

clinically suspected probability of a deep subcortical periventricular tumor growth ventriculography, as a rule, is not indicated because of the possibility of a vigorous general reaction in the presence of a tumor which is inoperable. In such cases, on the EEG usually only an individual focus of pathological electrical activity may be detected with a degree of expression of those general changes which are characteristic of signs of increased pressure and which characterize the rapidly growing immature glioblastomas.

Only through a comparison of the course of the neurological syndrome, the EEG data and results of the examination of the spinal fluid can the problem be decided about the possibility of using ventriculography (in individual cases -- pneumoencephalography). In the presence of xanthochromia, considerable changes in the spinal fluid, in the presence of electroencephalographic data of a rapidly growing deep tumor and corresponding features of a neurological syndrome ventriculography or pneumoencephalography in deep subcortical tumors of a periventricular character should be considered contraindicated. This is confirmed by numerous observations.

Bilateral arteriography in such cases is less dangerous.

It should be performed against the background of dehydration measures, even if it gives only a temporary effect.

Clinical study of the considerable number of patients with cerebral gliomas, investigated from many angles, who have been studied and subjected to operations, as well as the study of autopsy data permit us to draw a generalizing conclusion as to the modern trends and possibilities of precise preoperative diagnosis.

Such a diagnosis is made and substantiated by the consistent use of all modern methods of examination adopted in every individual case, from the very beginning being directed by an analysis of the neurological syndrome dynamically, including a study of ophthalmological and otoneurological data, like the results of investigation of the condition of central regulation of the cardiovascular system and metabolism. In neurosurgery successful use may be made of roentgenological and spinal fluid studies, the data of electroencephalographic recordings, arteriography, and where indicated, the data of ventriculography or pneumoencephalography. Only consistent use of a number of special and precision methods, supplementing the evaluation of the neurological syndrome, and a comparison of

the corresponding data make it possible with the greatest probability to make a precise preoparative diagnosis of cerebral gliomas.

The use of contrast methods only as a basis for the diagnosis, which has been successfully used by many neurosurgeons for meningiomas, is fraught with considerable dangers and errors of differentiation in gliomas.

Different courses of gliomas and glioblastomas, considerable differences in their clinical manifestations, in the use of diagnostic methods, the different manner of solving problems of operative indications in surgical tactics in the three principal focal categories of gliomas -- the cortical-subcortical, subcortical in the area of the centrum ovale and deep periventricular -- require the elaboration of methods of a most precise diagnosis which may be determined only through a complicated combination of examinations. The difficult chapter of neurosurgery of gliomas may be developed along this route with less danger and with fewer losses.

Received for publication

19 December 1959

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